

UNUSUAL AND MEMORABLE

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Case number 34: Relapse of polyarteritis nodosa presenting as isolated and localised lower limb periostitis

A 59 year old woman presented with progressively increasing pain and swelling over the right lower limb that had started at the end of 2001. No systemic manifestations such as fever, weight loss, arthralgia, myalgia, or rash were present. Upon examination we noticed a painful swelling localised over the middle one third of the right tibia without cutaneous alterations. There were no other pathological clinical signs. At that time, she was taking no drugs.

Her medical history disclosed systemic polyarteritis nodosa (PAN) in 1991, at which time she presented with pain and swelling at the knees and ankles as well as cutaneous nodules over the medial side of the left ankle. This clinical presentation, together with the inflammatory biology, the absence of autoantibodies, the presence of radiological and scintigraphic periostitis, and the finding of microaneurysms upon arteriography, led to the diagnosis of PAN. She was successfully treated with corticosteroids over a period of 12 months. Upon discontinuation of the steroid treatment, she remained in remission.

For further investigation of her recent problem of localised tibial pain and swelling, she was admitted to hospital. Blood examination showed an erythrocyte sedimentation rate of

38 mm/1st h and C reactive protein of 77 mg/l. A peripheral blood count, blood chemistry, liver function tests, renal function tests, and complement C3 and C4 levels were all within a normal range. Rheumatoid factor, antinuclear factor, antineutrophil cytoplasmic antibodies, and antiperinuclear factor were negative. Urine sediment was normal. Hepatitis B and C serology were negative. Radiology of the right tibia showed a periosteal reaction (fig 1A). Bone scintigraphy demonstrated localised tracer hypercapture over the symptomatic zone (fig 1B). A nuclear magnetic resonance scan of the right tibia disclosed a periosteal reaction suggestive of an inflammatory or infectious process (periostitis-osteomyelitis) (figs 1C and D). The differential diagnosis included a stress fracture, a bone tumour, or a relapse vasculitis. Arteriography of the truncus coeliacus showed normal findings. Biopsy specimens of the periost were obtained surgically and showed necrotising vasculitis compatible with PAN (figs 1E and F). On the basis of this histopathology, a relapse of PAN presenting as an isolated and localised periostitis of the right tibia was diagnosed. Oral prednisone treatment was restarted with good clinical response.

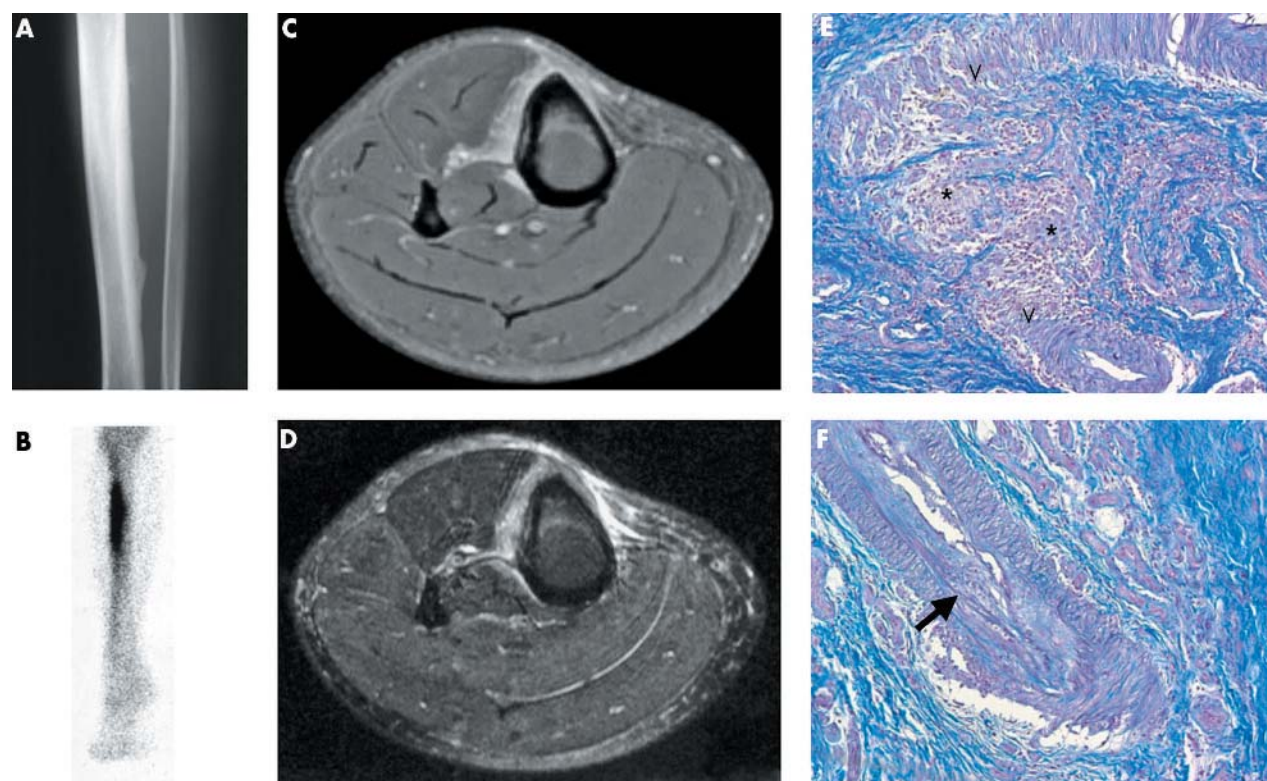


Figure 1 (A) Radiology of the right tibia, showing a periosteal reaction. (B) The delayed anterior and posterior bone scan image shows an intense elongated hot spot at the lateral border of the right tibia. (C, D) A nuclear magnetic resonance scan (D: after gadolinium) of the right tibia showed a periosteal reaction bordering the anterior and lateral part of the middle one third, with infiltration and inflammation of the weak tissues suggestive of an inflammatory/infectious process (periostitis-osteomyelitis). (E) In the thickened periosteal fibrous tissue a curved vessel (V) is found, the wall of which is segmentally infiltrated by chronic inflammatory cells (*). Trichromous stain, original magnification $\times 400$. (F) Middle sized artery, disclosing segmental inflammatory changes (arrow). Trichromous stain, original magnification $\times 400$.

Cases of periostitis in patients with PAN (often localised PAN) have been reported,¹⁻⁷ although mostly in older publications. The periosteal reaction is most often found in the lower limbs,¹⁻⁶ and may or may not be accompanied by cutaneous lesions.¹⁻⁶ Commonly, laboratory tests show mild to moderate inflammation. The current case report describes a relapse of systemic PAN, presenting only with lower limb pain and swelling and periosteal new bone formation. The blood examination showed moderate inflammation. Imaging raised the possibility of a bone tumour with periosteal reaction, which could also match the clinical picture. We therefore decided to perform a biopsy, which confirmed the diagnosis of PAN. The case report illustrates that vasculitis should also be considered in isolated and localised periostitis.

**P Carron, I E A Hoffman, L De Rycke, I Peene, E M Veys,
F De Keyser**
Department of Rheumatology, Ghent University Hospital, Belgium

W Bauters
Department of Radiology, Ghent University Hospital, Belgium

M Praet
Department of Pathology, Ghent University Hospital, Belgium

Correspondence to: Dr I Hoffman, Department of Rheumatology, Ghent University Hospital, De Pintelaan 185, 9000 Gent, Belgium; Ise.Hoffman@UGent.be

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